

Creutzfeldt-Jakob Disease (CJD)

What is CJD?

Creutzfeldt-Jakob disease (pronounced “kroits-felt-yah-cub”; CJD) is a rare brain disorder – only a few cases are diagnosed in the US each year. Many scientists believe that CJD is caused by a protein (called a “prion”) that destroys normal brain cells.

Who gets CJD?

Almost all cases occur in people over 45 years of age. In about 85% of patients, CJD occurs for no known reason. A smaller percentage of patients (5-15%) inherit the disease. In a few cases, people who received infected tissue during surgical procedures (corneal transplants, surgery on the brain or its coverings) have developed CJD. A different form of the disease (vCJD) is described at the end of this factsheet and is associated with eating meat from infected cows.

How is CJD spread?

Contact between people does not present a risk for transmission for CJD. The disease cannot be spread through the air or by touching someone with CJD.

CJD could be spread by certain medical procedures. Prions are very difficult to destroy by heat or chemicals, so patients could be infected if surgical instruments have not been sterilized adequately after use on a CJD patient. Hospitals are very aware of the need to use disposable equipment or take extra precautions to sterilize surgical instruments so all patients are protected against infection with prions.

What are the symptoms of CJD?

Early symptoms may include forgetfulness, behavior changes, and loss of coordination. Patients with CJD eventually lose the ability to talk, walk, and take care of themselves. The disease progresses rapidly and most patients with CJD die within a year of diagnosis.

How soon after exposure do symptoms appear?

It can take from 15 months to 30 years or more for symptoms to appear. Most of the time, there is no exposure to pinpoint. Rather, it is thought that something happens to make a normal prion change to a form that can cause disease, with no environmental source of infection.

How is CJD diagnosed?

There is currently no simple test for CJD. Although doctors may do a lot of tests to look for a cause of a patient’s illness, the only definite test for CJD is brain biopsy (removing a small piece of tissue from the patient’s brain for testing) or examination of the brain after death.

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What is the treatment for CJD?

Currently, there is no specific medication or cure for CJD. Care involves relieving symptoms and making the patient as comfortable as possible.

How can CJD be prevented?

People with CJD or people at increased risk should never donate blood, tissues, or organs.

Caregivers of CJD patients should use good hygiene, including:

- Wash hands and exposed skin before eating, drinking, or smoking. This is a good habit to practice, even though contact with intact skin of a CJD patient does not spread the disease.
- Cover cuts and abrasions with waterproof bandages.
- Wear surgical gloves when handling a patient's tissues and fluids or dressing a patient's wounds.
- Avoid getting cut or stuck with items (e.g., needles) that may be contaminated with body fluids.
- Use face protection if there is a risk of exposure to the patient's blood or body fluids.
- If a CJD patient's body fluids gets on someone else's skin, then wash the affected area with detergent, rinse well with warm water (avoid scrubbing), rinse the site with a 1:9 dilution of bleach for 1 minute, and then rinse again with water. If a CJD patient's body fluids get into someone else's eye or mucous membranes, then rinse well with saline or tap water.
- Dispose of all clinical waste (e.g., bandages) properly.

What is the relationship of CJD to “Mad Cow Disease”?

The disease that is referred to as “Mad Cow Disease” is a disease that occurs in cattle and is technically called bovine spongiform encephalopathy (BSE). Some people who have eaten meat from cattle with BSE have gotten a disease that looks like CJD but is caused by a different prion. This disease has been named ‘variant CJD’ (vCJD) and has been diagnosed in about 200 people in the world. In the US, only three cases of vCJD have been diagnosed; all in people who grew up and were exposed to BSE outside the US. The US Department of Agriculture has developed measures to screen cattle so BSE-infected animals are excluded from the food chain and their tissues cannot infect other animals or humans.

How can I get more information about CJD?

- 1) If you have concerns about disease, contact your healthcare provider.
- 2) Call your local health department. A directory of local health departments is located at <http://www.vdh.virginia.gov/LHD/index.htm>.
- 3) Visit the Centers for Disease Control and Prevention website at <http://www.cdc.gov/ncidod/dvrd/cjd/>.